

Serial Number: 091993, 241

CRF Processing Date: 12/6/01

Edited by: DC

Verified by: DC

(STIC sta

ENTERED

- ☐ Changed a file from non-ASCII to ASCII
- ☐ Changed the margins in cases where the sequence text was "wrapped" down to the next line.
- ☐ Edited a format error in the Current Application Data section, specifically
- ☐ Edited the Current Application Data section with the actual current number. The number inputted by the applicant was ☐ the prior application data; or ☐ other
- ☐ Added the mandatory heading and subheadings for "Current Application Data".
- ☐ Edited the "Number of Sequences" field. The applicant spelled out a number instead of using an integer.
- ☐ Changed the spelling of a mandatory field (the headings or subheadings), specifically:
- ☐ Corrected the SEQ ID NO when obviously incorrect. The sequence numbers that were edited were:
- ☐ Inserted or corrected a nucleic number at the end of a nucleic line. SEQ ID NO's edited:
- ☐ Corrected subheading placement. All responses must be on the same line as each subheading. If the applicant placed a response below the subheading, this was moved to its appropriate place.
- ☐ Inserted colons after headings/subheadings. Headings edited included:
- ☐ Deleted extra, invalid, headings used by an applicant, specifically:
- ☒ Deleted: ☒ non-ASCII "garbage" at the beginning/end of files; ☐ secretary initials/filename at end of file; ☐ page numbers throughout text; ☐ other invalid text, such as
- ☐ Inserted mandatory headings, specifically:
- ☐ Corrected an obvious error: in the response, specifically:
- ☐ Edited identifiers where upper case is used but lower case is required, or vice versa.
- ☐ Corrected an error in the Number of Sequences field, specifically:
- ☐ A "Hard Page Break" code was inserted by the applicant. All occurrences had to be deleted.
- ☐ Deleted *ending* stop codon in amino acid sequences and adjusted the "(A)Length:" field accordingly (error due to a PatentIn bug). Sequences corrected:
- ☐ Other:

*Examiner: The above corrections must be communicated to the applicant in the first Office Action. DO NOT send a copy of this form.

OIPE

RAW SEQUENCE LISTING

PATENT APPLICATION: US/09/993,241

DATE: 12/06/2001

TIME: 15:50:23

Input Set : D:\008000051CNUS01.txt

Output Set: N:\CRF3\12062001\I993241.raw

**Does Not Comply
Corrected Diskette Needed**

4 <110> APPLICANT: KAKKIS, EMIL D.
 6 <120> TITLE OF INVENTION: METHODS FOR TREATING DISEASES CAUSED BY DEFICIENCIES OF
 7 RECOMBINANT ALPHA-L-IDURONDINASE
 9 <130> FILE REFERENCE: 008000051CNUS01
 C--> 11 <140> CURRENT APPLICATION NUMBER: US/09/993,241
 12 <141> CURRENT FILING DATE: 2001-11-13
 14 <150> PRIOR APPLICATION NUMBER: 09/711,205
 15 <151> PRIOR FILING DATE: 2000-11-09
 17 <150> PRIOR APPLICATION NUMBER: 09/439,923
 18 <151> PRIOR FILING DATE: 1999-11-12
 20 <160> NUMBER OF SEQ ID NOS: 2
 22 <170> SOFTWARE: FastSEQ for Windows Version 3.0

ERRORED SEQUENCES

273 <210> SEQ ID NO: 2
 274 <211> LENGTH: 650
 275 <212> TYPE: PRT
 276 <213> ORGANISM: Homo sapiens
 278 <400> SEQUENCE: 2
 279 Met Arg Pro Leu Arg Pro Arg Ala Ala Leu Leu Ala Leu Leu Ala Ser
 280 1 5 10 15
 281 Leu Leu Ala Ala Pro Pro Val Ala Pro Ala Glu Ala Pro His Leu Val
 282 20 25 30
 283 His Val Asp Ala Ala Arg Ala Leu Trp Pro Leu Arg Arg Phe Trp Arg
 284 35 40 45
 285 Ser Thr Gly Phe Cys Pro Pro Leu Pro His Ser Gln Ala Asp Gln Tyr
 286 50 55 60
 287 Val Leu Ser Trp Asp Gln Gln Leu Asn Leu Ala Tyr Val Gly Ala Val
 288 65 70 75 80
 289 Pro His Arg Gly Ile Lys Gln Val Arg Thr His Trp Leu Leu Glu Leu
 290 85 90 95
 291 Val Thr Thr Arg Gly Ser Thr Gly Arg Gly Leu Ser Tyr Asn Phe Thr
 292 100 105 110
 293 His Leu Asp Gly Tyr Leu Asp Leu Leu Arg Glu Asn Gln Leu Gly Phe
 294 115 120 125
 295 Glu Leu Met Gly Ser Ala Ser Gly His Phe Thr Asp Phe Glu Asp Lys
 296 130 135 140
 297 Gln Gln Val Phe Glu Trp Lys Asp Leu Val Ser Ser Leu Ala Arg Arg
 298 145 150 155 160
 299 Tyr Ile Gly Arg Tyr Gly Leu Ala His Val Ser Lys Trp Asn Phe Glu
 300 165 170 175
 301 Thr Trp Asn Glu Pro Asp His His Asp Phe Asp Asn Val Ser Met Thr
 302 180 185 190
 303 Met Gln Gly Phe Leu Asn Tyr Tyr Asp Ala Cys Ser Glu Gly Leu Arg
 304 195 200 205

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```

305  Ala Ala Ser Pro Ala Leu Arg Leu Gly Gly Pro Gly Asp Ser Phe His
306      210                      215                      220
307  Thr Pro Pro Arg Ser Pro Leu Ser Trp Gly Leu Leu Arg His Cys His
308      225                      230                      235                      240
309  Asp Gly Thr Asn Phe Phe Thr Gly Glu Ala Gly Val Arg Leu Asp Tyr
310                      245                      250                      255
311  Ile Ser Leu His Arg Lys Gly Ala Arg Ser Ser Ile Ser Ile Leu Glu
312                      260                      265                      270
313  Gln Glu Lys Val Val Ala Gln Gln Ile Arg Gln Leu Phe Pro Lys Phe
314                      275                      280                      285
315  Ala Asp Thr Pro Ile Tyr Asn Asp Glu Ala Asp Pro Leu Val Gly Trp
316      290                      295                      300
317  Ser Leu Pro Gln Pro Trp Arg Ala Asp Val Thr Tyr Ala Ala Met Val
318      305                      310                      315                      320
319  Val Lys Val Ile Ala Gln His Gln Asn Leu Leu Leu Ala Asn Thr Thr
320                      325                      330                      335
321  Ser Ala Phe Pro Tyr Ala Leu Leu Ser Asn Asp Asn Ala Phe Leu Ser
322                      340                      345                      350
323  Tyr His Pro His Pro Phe Ala Gln Arg Thr Leu Thr Ala Arg Phe Gln
324                      355                      360                      365
325  Val Asn Asn Thr Arg Pro Pro His Val Gln Leu Leu Arg Lys Pro Val
326      370                      375                      380
327  Leu Thr Ala Met Gly Leu Leu Ala Leu Leu Asp Glu Glu Gln Leu Trp
328      385                      390                      395                      400
329  Ala Glu Val Ser Gln Ala Gly Thr Val Leu Asp Ser Asn His Thr Val
330                      405                      410                      415
331  Gly Val Leu Ala Ser Ala His Arg Pro Gln Gly Pro Ala Asp Ala Trp
332                      420                      425                      430
333  Arg Ala Ala Val Leu Ile Tyr Ala Ser Asp Asp Thr Arg Ala His Pro
334                      435                      440                      445
335  Asn Arg Ser Val Ala Val Thr Leu Arg Leu Arg Gly Val Pro Pro Gly
336      450                      455                      460
337  Pro Gly Leu Val Tyr Val Thr Arg Tyr Leu Asp Asn Gly Leu Cys Ser
338      465                      470                      475                      480
339  Pro Asp Gly Glu Trp Arg Arg Leu Gly Arg Pro Val Phe Pro Thr Ala
340                      485                      490                      495
341  Glu Gln Phe Arg Arg Arg Ala Ala Glu Asp Pro Val Ala Ala Ala Pro
342                      500                      505                      510
343  Arg Pro Leu Pro Ala Gly Gly Arg Leu Arg Leu Arg Pro Ala Leu Arg
344                      515                      520                      525
345  Leu Pro Ser Leu Leu Leu Val His Val Cys Ala Arg Pro Glu Lys Pro
346      530                      535                      540
347  Pro Gly Gln Val Thr Arg Leu Arg Ala Leu Pro Leu Thr Gln Gly Gln
348      545                      550                      555                      560
349  Leu Val Leu Val Trp Ser Asp Glu His Val Gly Ser Lys Cys Leu Trp
350                      565                      570                      575
351  Thr Tyr Glu Ile Gln Phe Ser Gln Asp Gly Lys Ala Tyr Thr Pro Val
352                      580                      585                      590
353  Ser Arg Lys Pro Ser Thr Phe Asn Leu Phe Val Phe Ser Pro Asp Thr

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Output Set: N:\CRF3\12062001\I993241.raw

```

354          595          600          605
355 Gly Ala Val Ser Gly Ser Tyr Arg Val Arg Ala Leu Asp Tyr Trp Ala
356      610          615          620
357 Arg Pro Gly Pro Phe Ser Asp Pro Val Pro Tyr Leu Glu Val Pro Val
358 625          630          635          640
359 Pro Arg Gly Pro Pro Ser Pro Gly Asn Pro
360          645          650
E--> 361 1

```

VERIFICATION SUMMARY

PATENT APPLICATION: US/09/993,241

DATE: 12/06/2001

TIME: 15:50:24

Input Set : D:\008000051CNUS01.txt

Output Set: N:\CRF3\12062001\I993241.raw

L:11 M:270 C: Current Application Number differs, Replaced Current Application Number
L:361 M:332 E: (32) Invalid/Missing Amino Acid Numbering, SEQ ID:2